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Picturing the Experience of Living With Myotonic Dystrophy (DM1): A Qualitative Exploration Using Photovoice

Kori A. LaDonna, Shannon L. Venance

ABSTRACT

Background: Myotonic dystrophy presents with multisystemic complications, and there is a well-recognized myotonic dystrophy personality profile that is characterized by executive dysfunction, an avoidant personality, and impaired cognition. Understanding symptom impact on patients' lives is crucial for providing appropriate patient-centered care; however, much of the myotonic dystrophy literature reflects the biomedical model, and there is a paucity of articles exploring patient experience. **Objective:** The aim of this study was to use a novel research approach to explore the experiences of patients with myotonic dystrophy. **Methods:** Nine individuals participated in a qualitative study using the photovoice method. Photovoice uses the visual image to document participants' lives, and participants took pictures pertaining to living with myotonic dystrophy that stimulated individual and focus group interviews. We used content analysis to analyze the data; in turn, codes were collapsed into themes and categories. Findings were presented to participants to ensure resonance. **Results:** Participants took 0–40 photographs that depicted barriers and facilitators to living successfully with myotonic dystrophy. We identified two categories that include participants' challenges with everyday activities, their worries about the future, their grief for lost function and social opportunities, and their resilience and coping strategies. Participants also described their experiences using the photovoice method. **Conclusion:** Photovoice is a useful approach for conducting research in myotonic dystrophy. Participants were active research collaborators despite perceptions that individuals affected with myotonic dystrophy are apathetic. Our findings suggest that participants are concerned about symptom impact on reduced quality of life, not symptoms that clinicians preferentially monitor. Nurses, therefore, are essential for providing patient-centered, holistic care for patients' complex biopsychosocial needs. Research exploring current physician-led clinical care models is warranted.

Keywords: myotonic dystrophy, patient-centered care, photovoice, qualitative research

Myotonic dystrophy—the most common adult form of muscular dystrophy—is a chronic, progressive, and inherited neuromuscular condition presenting with multisystemic complications including distal muscle weakness, myotonia, hypersomnolence, early-onset cataracts, cardiac conduction abnormalities, and slurred speech and swallowing problems (Harper, 2001). The complex needs of patients with

myotonic dystrophy—including their cognitive and behavioral impairments—may complicate patient-centered care provision. In particular, patients' lower educational attainment, problems with executive function, and avoidant personality traits may make it difficult for them to take risks, make friends, or participate in new activities (Delaporte, 1998; Gagnon, Mathieu, & Noreau, 2007; Meola et al., 2003; Sistiaga et al., 2010); consequently, individuals with myotonic dystrophy may be apathetic and disinterested in their health (Meola & Sansone, 2007).

Despite these challenges, it is essential that patients' values and experiences drive clinical care. Clinicians are concerned about the potential for cardiac, respiratory, or swallowing abnormalities to cause morbidity or sudden death; yet, research exploring patients' perspectives suggests that weakness, fatigue, and myotonia have a greater impact on patients' quality of life (Boström & Ahlström, 2004; Cup et al., 2011; Gagnon et al., 2007; Heatwole et al., 2012; Nätterlund, Sjöden, & Ahlström, 2001). These symptoms often challenge affected individuals' ability to socialize, complete household chores, or engage in employment or educational

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opportunities. Progressive physical symptoms—coupled with patients' difficulties with motivation and planning—may impact patients' personal relationships (Cup et al., 2011). Consequently, patients may experience significant disruption and decreased satisfaction with employment and social recreation (Gagnon et al., 2007); in turn, impaired mental and physical function and reduced quality of life may result (Laberge et al., 2013).

Despite evidence that clinicians and patients may have different concerns and goals, much of the current myotonic dystrophy literature is written from a biomedical perspective, and relatively few qualitative studies explore the perspectives of patients with muscular dystrophy (LaDonna, Koopman, Ray, & Venance, in press; Boström & Ahlström, 2005; Boström & Ahlström, 2004; Boström, Ahlström, & Sunvisson, 2006; Cup et al., 2011; Faulkner & Kingston, 1998; Heatwole et al., 2012; Nätterlund et al., 2001). These studies use a range of methods including content analysis and phenomenology to explore questions pertaining to living with muscular dystrophy; few of these studies represent the perspectives of North American patients, and individuals with myotonic dystrophy are a small proportion of the overall sample. We believe that individuals living with myotonic dystrophy have valuable insights into their condition and life experiences that can inform their health management and that their voices should be more widely distributed in the literature. To address this, we used an innovative qualitative research method called photovoice (Wang & Burris, 1994, 1997) to explore the experience of living with myotonic dystrophy.

Photovoice is rooted in participatory action research and was developed to study populations with low literacy residing outside the traditional power structure (Wang & Burris, 1994). "Participatory Action Research differs from most other approaches to public health research because it is based on reflection, data collection, and action that aims to improve health and reduce health inequities through involving the people who, in turn, take actions to improve their own health" (Baum, MacDougall, & Smith, 2006). Although we do not believe that individuals with myotonic dystrophy are "powerless," we argue that their unique physical and emotional challenges, coupled with the power imbalance inherent in some medical encounters (Goodyear-Smith & Buetow, 2001), warrant a strategy that utilizes their strengths and experiences. Photovoice uses photography to augment traditional interviews by offering participants a creative way to communicate their expertise, experiences, knowledge, and needs. In an effort to educate or enact change, participants may choose to disseminate their photographs and experiences to "stakeholders" like clinicians or patient advocacy groups (Wang & Burris, 1994, 1997). Photovoice

Using photovoice, a unique qualitative research approach, this work explores the experiences of patients living with myotonic dystrophy.

has been used with individuals with intellectual impairment (Jurkowski & Paul-Ward, 2007), acquired brain injury (Lorenz & Kolb, 2009), stroke (Levin et al., 2007), dementia (Genoe & Dupuis, 2013), and Alzheimer disease (Wiersma, 2011).

The purposes of this study were to add voices of patients with myotonic dystrophy to the literature and to explore patients' experiences living with a chronic and progressive neurological disease. We asked participants to reflect on the impact of symptoms on their daily lives and to consider the barriers and facilitators to living successfully with myotonic dystrophy. We also sought to engage individuals as research collaborators; the literature describes that patients with myotonic dystrophy may be apathetic or cognitively impaired, so we also asked the question: Is photovoice an effective methodology for exploring the perspectives of individuals living with myotonic dystrophy?

Methods

Participants with mild-to-moderate adult-onset myotonic dystrophy (by physician report) attending an academic neuromuscular clinic were invited to participate. We purposively selected individuals who were able to provide informed consent and comply with study procedures; nine participants (four women) consented, and each was given a pseudonym to ensure confidentiality (Table 1).

We used a typical photovoice study consisting of a camera orientation session, an individual interview, and a focus group (Wang & Burris, 1997). Participants attended a camera orientation session in which they (a) discussed the ethics of picture taking, (b) were given a digital camera and instructed about its use, and (c) were asked to "take pictures of what it is like to live with myotonic dystrophy." In addition, participants were asked to take pictures of people or things that either hindered or facilitated living successfully with myotonic dystrophy. Instructions were purposely kept vague to avoid the potential for researchers to influence picture taking. Participants were instructed that they had to seek written consent from each person they wished to photograph. Researchers asked questions to check the participants' comprehension and

TABLE 1. Photovoice Participants

Patty	Patty is in her 50s and has mild myotonic dystrophy. Patty asked her husband to assist her with data collection; together, they took four pictures. She completed her individual interview but did not attend her scheduled focus group session.
Marjorie	Marjorie is in her 70s and has mild–moderate myotonic dystrophy. Marjorie’s husband helped her take pictures, and he attended her individual interview. They discussed 13 photographs. Although Marjorie was an enthusiastic research participant, she withdrew from the study because of fatigue.
Tim	Tim is in his 40s and has moderate myotonic dystrophy. He took 15 pictures and participated in focus group 1.
Frank	Frank is in his 40s and is moderately affected. He took 40 pictures and participated in focus group 1.
Joe	Joe is in his 40s and is mildly–moderately affected. Joe took 11 pictures and participated in focus group 2.
Max	Max is in his 40s and has moderate myotonic dystrophy. He was unable to take pictures because he had difficulty using the camera. Instead, his interview was structured around the images he would have taken. Max participated in focus group 2.
Jenny	Jenny is in her 40s and has mild myotonic dystrophy. Jenny chose the subject matter of her 28 images, but her partner took the pictures on her behalf. Both Jenny and her partner participated in the individual interview session, and Jenny was a member of focus group 2.
Peter	Peter is in his 50s and has moderate myotonic dystrophy. Peter stated that he had difficulty using the camera; consequently, he was only able to capture one image. Peter’s individual interview was used as a member check of the preliminary findings.
Meg	Meg is in her 40s and is mildly affected. Meg took five photographs, and her individual interview was used as a member check of the preliminary findings.

understanding of the task and instructions, and participants were given the opportunity to practice taking pictures.

After the camera orientation session, participants had 2–3 weeks to take pictures and then returned for an individual interview. Participants were asked to describe each of their photographs; in turn, their narratives directed the content and flow of the interviews. To further probe participants’ responses, the researchers prepared general questions about their medical history, their symptoms, and the impact of myotonic dystrophy on their daily activities. We also asked participants to share their opinions about study participation and the photovoice method. Participants were then invited to choose two-to-three photographs to share during a focus group.

All nine participants completed the camera orientation session and an individual interview. The first seven participants were scheduled to participate in a focus group, and two focus groups were held with two and three participants each. Two individuals declined participation; one participant felt her fatigue precluded focus group participation, and one participant was lost to follow-up. All data were retained and analyzed. After the focus groups, two additional participants were recruited, and their photographs and individual interview data were used to verify preliminary themes (Table 1). All interviews were recorded and transcribed verbatim.

In keeping with the tenets of participatory action research and photovoice (Wang & Burris, 1994, 1997), participants collaborated with the iterative data collection

and analysis process. The first step in data collection and analysis began when participants chose what to photograph. Photographs were then selected, contextualized, and coded. Participants selected photographs to discuss and gave them meaning by describing what was captured and how and why they chose the subject matter (contextualization). We used content analysis to analyze the interview transcripts; in particular, a double-coding qualitative method was used to enhance the reliability of the data coding system (Miles, Huberman, & Saldana, 2014). We independently coded segments of the individual interview and focus group data using words or phrases that described participants’ actions or experiences. We held regular meetings to discuss the coding, and a preliminary list of themes was developed by consensus. Data collection and analysis were iterative, and preliminary themes were discussed during subsequent interviews to ensure that the findings resonated with participants’ experiences (Bradbury-Jones, Irvine, & Sambrook, 2010). No new themes emerged during the final two interviews, and recruitment ceased when we determined that the collected data were sufficient for exploring our research questions. Although additional participants might have generated new insights, we determined that our themes and categories provided a robust exploration of participants’ experiences of living with myotonic dystrophy. Transcripts were reexamined and then recoded using the finalized list of themes. Themes were then consolidated into categories. We kept a reflexive journal throughout the study to record general impressions of emerging

findings and to track theme and category development. Nvivo 10, a qualitative software program, was used to organize and manage the data.

This study complies with the Helsinki Declaration of 1975 and was approved by the Western University Research Ethics Board.

Results

Participants took 0–40 photographs that captured symptom impact on their activities and quality of life, their management strategies, and their sense of self. We identified seven themes that were consolidated into two categories titled “A Shifting Identity” and “Managing Limitations.” Participants described that their shifting identities caused them to lose their sense of self and to grieve for their lost abilities; however, they were able to find solutions to cope with their evolving needs. Participants also discussed their variable degrees of study participation; whereas some participants had difficulty following study procedures, others were able to problem-solve strategies to mitigate challenges. Regardless, all participants provided rich descriptions about their experiences living and coping with myotonic dystrophy.

Participating in Research

In general, participants were actively engaged in the project and stated that they enjoyed participating because it gave them the opportunity to share their experiences and socialize. Some participants were either the first in their family to be diagnosed or had never met anyone with myotonic dystrophy outside their immediate families. Consequently, the focus group was an opportunity for them to meet other affected individuals and discuss common experiences:

I meet a lot of people with MD (*muscular dystrophy*), but it's a big umbrella. So there's all different kinds. But I only know one person that has myotonic dystrophy and I'd like to meet other people that have myotonic dystrophy. Like males, females, different ages, and stuff like that. Maybe there'll be a friendship, you know? 'Cause we can talk and we understand what each other is going through, or something like that. (Tim)

There were, however, challenges associated with using an innovative qualitative research approach with this population. Transportation was problematic; two non-ambulatory participants (Jenny and Frank) lived approximately 2 hours from the study site. Whereas Jenny's caregiver was able to drive her, we had to arrange special transportation for Frank. As a result, Frank's individual and focus group session were conducted the same day, and he stated that the long study visit made him feel fatigued. In addition, Frank's

speech impairment rendered it difficult to understand and transcribe his comments. Two participants (Patty and Marjorie) did not complete the study; Marjorie withdrew citing fatigue, and Patty did not attend her scheduled focus group meeting. Finally, two participants (Max and Peter) had difficulty using the digital camera:

I was trying to...on Wednesday I was trying to take pictures of my bath seat and my walker. I put the camera on, I pushed it and it didn't go off, the flash stopped working. So...Well, I wasn't sure what to do, so I thought I'd just come down and tell you. Maybe I pushed the button that I shouldn't have, you know? (Peter)

Four individuals (Tim, Patty, Marjorie, and Jenny) facilitated their participation by asking a friend or care partner to help them take pictures. Together, Marjorie and her husband created a list of her challenges and most problematic symptoms and then brainstormed scenarios to photograph these limitations. Other care partners assisted with picture taking and/or participated in the individual interviews. All participants described their interest in participating in research in general—and this project in particular—because it gave them the opportunity to educate researchers and clinicians about their lives.

It gives them an idea of what it's like and learn that it's not all negative and that...like I say, you don't know what it's like unless you live it, but to take pictures you can see what my life is like. Not all negative, but not all positive either. Yeah, it's a good idea. (Frank)

A Shifting Identity

Participants described that their diagnosis caused an identity shift; that is, participants' symptoms altered their physical appearance and challenged their abilities to be employed, complete household chores, or participate in recreational activities. Consequently, progressive disability caused participants to lose their sense of self and grieve their presymptomatic identities.

A Changing Body

Participants' evolving physical appearance triggered their perception of a shifting identity; in particular, participants stated that their altered appearance and progressive functional decline challenged their self-confidence and their social participation. Participants commented that their changing bodies—including low muscle tone, drooping eyelids, premature balding, and facial atrophy—coupled with their need for assistive devices, significantly impacted their self-esteem.

Consequently, participants worried that they were unattractive and that their functional decline made them feel older than their chronological age:

I can't wear high heels' cause I fall of them! I gotta buy shoes with a low heel and that makes me feel so *stupid* because I feel like an old lady. You know, with a walker I feel like an old lady. I'm not—I'm 51—but that's not old to me you know? (Patty)

Even participants in stable and loving relationships experienced poor self-esteem; some participants were embarrassed not only by their physical appearance but also by impaired mobility and their propensity for falls: “It's not so much the scrapes on your hands and knees, it's the scrapes on your *dignity*...” (Meg). Participants felt judged and believed that members of the general public lacked empathy and treated people with disability unfairly. When asked to comment about her shifting identity, Jenny stated: “Thank God I knew people when I was still *okay*—walking around and stuff—because otherwise I think I'd probably be treated differently, being in a wheelchair, people not knowing. Personality doesn't change because you're in a wheelchair.” Similarly, other participants believed that members of the general public perceived them as slovenly or cognitively impaired, particularly if they had a speech impairment or fell in public: “I fall, they judge me as clumsy, and when I appear with mud on me because I've fallen, they assume I don't bother bathing regularly. So...that bothers me, the people that assume” (Meg).

The Challenges of Everyday Activities

Symptoms including weakness, gastrointestinal problems, and chronic respiratory failure impeded participants from fully participating in activities they once enjoyed. Participants with diarrhea were concerned about incontinence, and individuals with breathing difficulties found it cumbersome to travel with their BiPAP machines. Participants described that symptoms also impacted their ability to do activities usually taken for granted like bathing, walking a short distance, or climbing stairs: “I don't like stairs.... Walking down stairs is fine, but going up stairs...It's hard on my legs, eh? I go up two flights and I breathe hard.... Yeah, and I have to sit there and wait for a while. I sit there for a while, and then we'll go up two stories. I've got five flights of stairs to go up” (Peter). Limited finances made it difficult to participate in recreational activities, and watching television was the primary leisure activity for several participants. In addition, participants described difficulty exercising or opening doors (Fig 1) and bottles or jars, and the local environment was described as inaccessible to those with disabilities.

Narrow aisles, sidewalks in poor condition, and a lack of handicapped accessible entrances made it difficult to navigate these spaces:

I was going to take a picture of the mall because it's the only one that on the centre doors it has no buttons. It doesn't make sense because the bus, the Paratransit van, goes to a different door. That doesn't make sense to me because it should be on these two doors. (Frank)

Whereas myotonia, swallowing dysfunction (Fig 2), cardiac abnormalities, and cataracts minimally impacted participants' lives, fatigue was described as “a vicious enemy” (Marjorie) that exacerbated other symptoms. For example, Frank stated that his speech impairment worsened when he was tired, and excessive daytime somnolence caused participants to fall asleep at inappropriate times, making it difficult to work, complete chores, or socialize:

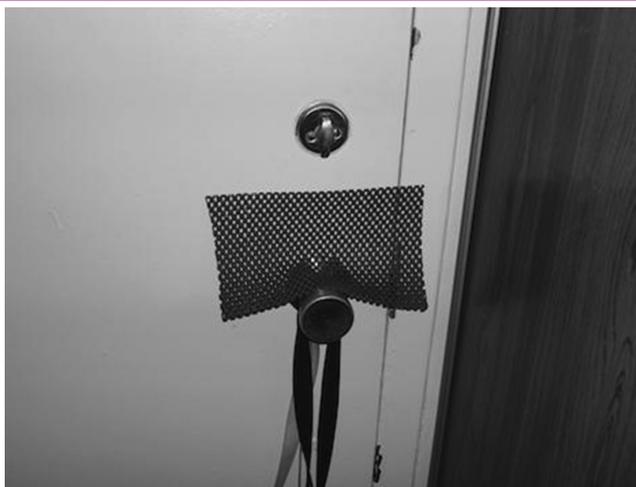
Falling asleep in the middle of conversations, it's embarrassing because I'll fall asleep in the middle of sentences, and it's not for lack of attention or lack of interest, it's just the myotonic dystrophy makes me tired. (Meg)

Loss and Grief

Participants' functional decline and physical appearance left them “constantly grieving” (Marjorie) their presymptomatic identity. Others described grieving for lost dreams, opportunities, and their previous physical abilities. Some participants had few social connections because their symptoms precluded full-time employment or participation in recreational or leisure activities. Consequently, participants' relationships were impacted by limited finances, fatigue, or a lack of motivation to be social. Tim and Frank perceived that their disabilities were off-putting to potential romantic partners, and some participants mourned their childless state because of infertility or because they feared transmitting the gene. Ultimately, participants grieved who they had been before their diagnosis and who they might have become had they not inherited myotonic dystrophy:

All through high school, I wanted to be a dancer. I used to take dance class and vocal class because I thought I was going to make it big and be discovered one day. There were times when I couldn't do stuff and they didn't understand why I couldn't do it. My flexibility was going and that was probably early signs of MD. Now, there's no way I could do that now, no way at all. I couldn't stand on a stage for long periods of time, dance or anything like that. There's no way. (Tim)

FIGURE 1 Improvising to Manage the Challenges of Everyday Activities: Using a Rubber Grip to Open a Door. “I find sometimes, especially if it’s cold, if I’m cold, they (hands) don’t always work, like when they seize up. And some door handles, the outside doors are easy, it’s just a push latch, it’s fine, which I like better.... But sometimes it’s just too damn hard.” (Joe)



Managing Limitations

Despite mourning their lost opportunities and progressive functional decline, participants were resilient and proactive about finding solutions to manage their limitations. Participants tempered their concerns about the future by adjusting their self-perceptions; that is, participants strategized solutions for mitigating their physical challenges and for finding purpose within their current abilities.

Desiring a Sense of Purpose

Participants expressed the need to feel valued and to be seen as contributing members of society: “Sometimes I’ll sort it (*laundry*) upstairs and that way at least they know I’m doing *something*...” (Patty). Many described desiring a sense of purpose and sought to maintain their independence and sense of control. Individuals took pride in their abilities and sought validation in volunteering or engaging in part-time

FIGURE 2 Making Swallowing Safer. “Well, that’s (swallowing) not good now.... But my jaw is not working well. I can’t chew.... I can’t eat beef, steak or, yeah, the chewing is a problem and I just get tired of chewing at the table and I stop.... You know, I manage to have ground meats, but even so, I don’t chew them. I just mush them around and swallow and I know that’s not good but that’s all I can do.” (Marjorie)



work, being proactive about their health, driving, and completing household chores. There was a clear sense that they tried to reframe their lost identity by focusing on their achievements and current abilities. Like Joe and Frank, Max described that he had once been an award-winning athlete; however, when asked about his current accomplishments, he quietly stated that he took pride in:

Staying alive, basically. Being able to live on my own and function as a human being to do what I do. I'm able to watch TV, listen to the radio, talk on the phone and try to keep my place presentable to anybody who's going to come over.

Finding Solutions

Participants described changing their expectations, creating new goals, and finding solutions for everyday challenges. In particular, individuals discovered tools or strategies to help them maintain their independence and functional ability while bolstering their self-esteem. These strategies included engaging in volunteer or paid employment, using mobility aids (Figure, available as Supplemental Digital Content 1 at <http://links.lww.com/JNN/A43>), taking medication for excessive fatigue, and facilitating household chores by using devices like rubber grips for opening jars or doors (Fig 1). Participants also adjusted their expectations about social and recreational outlets:

It's there, it's going to just get worse, but it hasn't really changed my lifestyle. Other than, yeah, less exercise, less sports.... But I mean participating, even kicking a ball around with the kids. *I can't do it.* Which is pissing me off, but you get over it, you know, and do something else. I can still play cards. (Joe)

Participants also coped by moving to accessible or community oriented housing, appreciating humor, and having pets. To preserve a “non-disabled” sense of self, some participants were selective about disclosing their diagnosis. For example, Tim's friends and family were aware of his condition, but he stated that he did not take his cane to work because he worried that his co-workers would treat him differently.

Family Dynamics

However, participants relied on family members or friends to assist them with completing tasks and maneuvering the local environment. Strong family, friend (Figure, available as Supplemental Digital Content 2 at <http://links.lww.com/JNN/A44>), and marital relationships seemed key to living successfully with myotonic

dystrophy. Individuals with a strong support system described having more coping strategies (particularly if there was financial stability or someone to help with chores) and feeling less isolated (particularly if they lived with their spouse or children). Overall, there was a sense that supportive family and community members eased the burden of disease and made individuals feel safe. When asked what information would be important for researchers and healthcare providers to know, Marjorie replied:

To fight depression. Because she's (*neurologist*) asked me about that. My doctor too. But I am not depressed because I have that man (*husband*) in my life. I couldn't be depressed.

Conversely, participants who lived alone, were unemployed, or lacked family support described feeling bored and isolated. Family or friends could exacerbate participants' feelings of isolation, disability, and dependence. Tim's gene-negative sister would not acknowledge his myotonic dystrophy, and some nonaffected family and friends did not understand the limitations imposed by fatigue and weakness. For example, Peter felt pressured to participate at a level that was discordant with his symptoms:

That's part of the myotonic dystrophy, it makes you tired, eh? Mom tells me I should be out longer. I say, Mom, you haven't got it. I'm *tired*. She says, “well, you shouldn't be tired.” “Well, I am, Mom.” You know, I'm almost 50 years old, I get tired.

What Will the Future Hold?

Participants were resilient despite worries about what the future might hold as their condition progressed. Many of these fears stemmed from having watched a family member decline; some participants had a “crystal ball” mentality and envisioned that total disability was imminent and inevitable: “Probably as my disease starts deteriorating. My hands—you know, pretty soon I won't be able to do anything and I'll go to a nursing home” (Peter). Meg worried about what would happen to her son with congenital myotonic dystrophy should she become incapacitated, and others were concerned about burdening family members with their care. In general, participants seemed more concerned about the effects of their decline on others, than for themselves.

Participants described problem-solving strategies to assuage their fears. In particular, participants described using bath seats and grab bars in the shower and putting spikes on their shoes during the winter to prevent falls. Moreover, some made plans for the future—albeit reluctantly—including financial plans and medical directives. For example, Marjorie stated

that she did not want a feeding tube should her dysphagia worsen. However, there was a variable amount of financial and supportive resources available to participants, and those with supportive care partners or paid employment seemed more secure about their future.

Others coped by not dwelling on having myotonic dystrophy and described that there were others living with far worse conditions or limitations: "So you do what you can, you do what you have to deal with, there's a lot of people who have a lot of bigger deals than this" (Meg). The primary coping strategy for most participants was to "just deal with it" and to take each day as it comes:

When it changes, you've got to make the change. That's all there is to it. You can't get it back. When it's gone, it's gone. That's the way this disease works. You have to learn to function with it. You've got to do what you can do with what you've got left. You do need assistance. You do need people around you who can help you when you need help. Don't be afraid to ask for help, that's what they're there for, utilize that. It may not be what you're used to but you have to make the change if you're going to get along with the disease. (Max)

Discussion

Shared decision making is the "pinnacle" of patient-centered care (Barry & Edgman-Levitan, 2012), and patients are increasingly collaborating with researchers to set clinical goals and research agendas (Schipper, Dauwerse, Hendriks, Leedekerken, & Abma, 2014; Teunissen, Visse, de Boer, & Abma, 2013). Recently, a mixed methods study assessing the research goals of patients with neuromuscular disease found that patients prioritize research that explores symptomatic management and medical care, their quality of life, and educational initiatives to raise awareness about neuromuscular disease (Nierse, Abma, Horemans, & van Engelen, 2013). There remains, however, a relative lack of patient-centered, qualitative research that explores the experiences of individuals living with myotonic dystrophy; consequently, there is little evidence that patients' voices are being heard or that their clinical and psychosocial needs are being met.

To address this, we successfully used a novel and innovative qualitative research approach that explores all of the patient-identified psychosocial research goals reported by Nierse et al. (2013). Although photovoice has been used with patients with a variety of chronic neurological conditions (Genoe & Dupuis, 2013; Levin et al., 2007; Lorenz & Kolb, 2009; Wiersma, 2011), it has not been used in myotonic dystrophy. We found photovoice to be a useful research method

for exploring the experiences of individuals living with myotonic dystrophy; our research participants captured a range of experiences including symptom impact on their quality of life, their concerns about the future, and their coping strategies. Although the qualitative approaches used in previous muscular dystrophy research provide rich data, we believe that photovoice is particularly beneficial for providing a unique perspective of patients' lives while giving them the opportunity to collaborate in research. Participants' photographs literally provide clinicians and researchers with a "picture" of patients' lives that would not be elucidated during a clinical encounter.

There were, however, limitations and challenges to using this innovative method with this population. We speculate that Max's and Peter's hand weakness or myotonia made it difficult for them to hold the camera and depress the shutter; in turn, they were unable to problem solve a solution for taking pictures. Symptoms including fatigue, speech impairments, mobility problems, and apathy may have further impacted participants' ability to comply with study procedures. However, although symptoms complicated participation, the benefits of photovoice far outweighed the challenges. We suggest that modifications or adaptations may be necessary to facilitate participation for some individuals. For example, although we gave participants the opportunity to practice taking pictures, we suggest that more time should be spent during the camera orientation session to clarify instructions and verify that participants are comfortable using the camera. It may also be advantageous to present participants with scenarios and then discuss potential problem-solving strategies should the camera fail to work or fatigue or mobility impairments hinder their ability to take pictures. Finally, we did not set out to include caregivers as research collaborators, but our research participants identified them as valuable for facilitating their study participation. Therefore, it may be helpful to include caregivers at the outset of photovoice projects exploring patients with complex needs. Despite challenges, participants were enthusiastic research collaborators who were candid about the impact of their progressive symptoms on their self-esteem, their social participation, and their relationships.

This study provides a rich exploration of the impact of myotonic dystrophy symptoms on patients' lives and illustrates patients' shifting identities, a concept that has not yet been fully articulated in the myotonic dystrophy literature. Our findings suggest that symptoms like dysphagia or cardiac abnormalities are not overly troubling for participants despite their propensity to cause sudden death (de Die-Smulders et al., 1998; Garrett, DuBose, Jackson, & Norman, 1969). Instead, participants were troubled by symptoms that

directly impacted their quality of life like weakness, fatigue, and their altered physical appearance. These findings resonate with other patients with muscular dystrophy (Boström & Ahlström, 2004; Cup et al., 2011; Gagnon et al., 2007; Heatwole et al., 2012; Nätterlund et al., 2001), and clinicians' and patients' divergent goals are well described in the neurological disease literature (Bonelli & Hofmann, 2004; Heesen, Köpke, Richter, & Kaspar, 2007; Martinez-Martin, Rodriguez-Blazquez, Kurtis, Chaudhuri, & NMSS Validation Group, 2011). A study of caregivers for individuals with myotonic dystrophy and dysphagia found that weakness and fatigue are more concerning to caregivers than swallowing dysfunction, despite the potential for choking to lead to aspiration pneumonia or sudden death. It is likely that the caregivers' concerns mirror those of their affected family members (LaDonna, Koopman, Ray, & Venance, in press).

Study participants also described the impact of myotonic dystrophy on their relationships and discussed their feelings of isolation and boredom. Participants articulated that the genetic and progressive nature of myotonic dystrophy caused them to mourn for lost abilities and opportunities. This sense of grief and loss permeated the photographs and interview transcripts, whether it was the death of a child with myotonic dystrophy or the progressive loss of function and social outlets. Grief resulting from being childless—because of infertility or fear of transmitting the gene—was echoed by participants in a qualitative study examining the experiences of 46 individuals living with a hereditary muscle disease (Boström & Ahlström, 2005). In addition, some of our study participants also expressed a sense of failure or self-blame for their infertility or for giving birth to a child affected by congenital myotonic dystrophy. Finally, while mourning the loss of the prediagnosis self is a common theme for individuals with muscular dystrophy—particularly in regard to embarrassment or self-consciousness about one's appearance (Boström & Ahlström, 2005)—the extent and complex nature of the grief of patients with myotonic dystrophy over their progressive functional decline has not yet been described. We posit that our participants experienced a cyclical pattern of loss: Disability caused the loss of independence through reduced recreational opportunities and employment. In turn, these losses caused financial and social restrictions that resulted in greater social isolation and loss of independence. Ultimately, these losses contributed to a decreased sense of self that was attributed to disability, thus perpetuating the cycle.

However, participants were resilient; their strategies for coping and maintaining independence were similar to those reported by other research participants and included doing chores or other activities at their

own pace or reframing their expectations for personal, social, or recreational goals (Boström & Ahlström, 2004). In this study, although participants reflected on their past accomplishments and their present limitations with some degree of sadness and frustration, they demonstrated resilience by focusing on their current achievements. Moreover, contrary to the established literature regarding the myotonic dystrophy personality profile (Delaporte, 1998; Meola et al., 2003), several of our participants showed initiative and resilience by putting their condition into perspective and taking it “day-by-day,” having a good knowledge base about their condition and an appreciation about prognosis, finding solutions to challenges, and being attentive and engaged study participants. Three participants were employed (paid or volunteer), and two were advocates for their condition; one is a leader in the myotonic dystrophy community, and another chose to attend a first-year Masters of Occupational Therapy course to discuss five of his pictures and answer questions about living with myotonic dystrophy. The latter was in keeping with the tenets of photovoice to disseminate information to “stakeholders.” Our results showing participants' advocacy, perseverance, and insight into their condition are significant in light of the current literature that focuses on the apathy and limited cognitive capacity affecting those with myotonic dystrophy.

Recommendations/Future Directions

Results from this study suggest that our study participants with mild-to-moderate myotonic dystrophy are proactive about finding solutions for challenges. Their photographs offer a window into participants' lives that might not be accessible during a clinic visit. Therefore, the participants' photographs of barriers and facilitators to living successfully with myotonic dystrophy might inform clinical recommendations, particularly suggestions for mobility aids or devices for assisting with daily tasks. It may also be helpful to use the photographs to create handouts or a poster of assistive devices or mobility aids for patients to use as a reference guide.

This study identified a need for an in-depth, examination of facets of living with myotonic dystrophy including isolation, grief, and opportunities for social and recreational pursuits; qualitative research is well suited to these inquiries. Finally, because participants in this study considered symptoms that clinicians provide rigorous surveillance for (e.g., dysphagia or cardiac abnormalities) to be minimally impactful, it would be valuable to interview healthcare providers and patients to ascertain which symptoms are of personal and clinical significance.

This finding also underscores the importance of, and need for, a multidisciplinary team that is situated

in either a neuromuscular, rehabilitation, or family practice clinic. To holistically address patients' complex needs, it is imperative that the team includes physicians, nurses, occupational and physical therapists, speech pathologists, and social workers. Nurses are educated specifically to collaborate with patients and their family members to address physical and emotional issues; in turn, nurses can help patients and caregivers navigate the healthcare system and obtain appropriate care in the hospital or in the community (W. J. Koopman, personal communication, September 19, 2014). Nurses, therefore, are well suited to provide holistic, patient-centered care for the complex needs of patients with myotonic dystrophy. In particular, nurses have the skill set to address patients' and caregivers' educational and psychosocial concerns while monitoring their symptoms and treating complications. Research suggests that patients who attend nurse-led chronic disease clinics have better self-care behaviors, improved outcomes, and greater satisfaction with their care (Hill, 1997; Stromberg et al., 2003). Although researchers have proposed a myotonic dystrophy management model and a nurse-led integrated clinical care pathway (Chouinard et al., 2009; Gagnon et al., 2007), these have not yet been systematically studied or implemented across clinical sites. More research is therefore needed to explore health care professionals and patients' perspectives of—and expectations for—a patient-centered approach to clinical care.

Limitations

This research study explored the experiences of nine individuals living with myotonic dystrophy and is therefore not generalizable to a wider population of patients. However, the results may resonate with patients in other clinical settings, and findings from this article may be useful for generating new research questions.

Conclusion

Photovoice offers an in-depth exploration of patients' experiences that may impact clinical care. Although it is important to consider the unique challenges presented by those living with myotonic dystrophy, photovoice is an informative and appropriate research method for exploring their lived experience. In particular, the participants' photographs and stories offer insight into patients' lives that would not typically be ascertained during a clinical encounter. Furthermore, incorporating patients' knowledge about myotonic dystrophy and their strategies for disease management may be useful for guiding educational initiatives and clinical recommendations. Finally, we argue that an awareness of—and appreciation for—patients' lived experience contributes to better patient-centered care and that photovoice is useful for uncovering patients' illness

experiences and generating research questions. Nurses may be particularly well suited to addressing the complex biopsychosocial needs of patients with myotonic dystrophy, and we propose that current physician-led myotonic dystrophy clinical care models warrant further investigation.

References

- Barry, M. J., & Edgman-Levitan, S. (2012). Shared decision making—The pinnacle of patient-centered care. *New England Journal of Medicine*, *366*, 780–781.
- Baum, F., MacDougall, C., & Smith, D. (2006). Participatory action research. *Journal of Epidemiology and Community Health*, *60*(10), 854–857.
- Bonelli, R. M., & Hofmann, P. (2004). A review of the treatment options for Huntington's disease. *Expert Opinion on Pharmacotherapy*, *5*, 767–776.
- Boström, K., & Ahlström, G. (2004). Living with a chronic deteriorating disease: The trajectory with muscular dystrophy over ten years. *Disability and Rehabilitation*, *26*(23), 1388–1398.
- Boström, K., & Ahlström, G. (2005). Living with a hereditary disease: Persons with muscular dystrophy and their next of kin. *American Journal of Medical Genetics, Part A*, *136*(1), 17–24.
- Boström, K., Ahlström, G., & Sunvisson, H. (2006). Being the next of kin of an adult person with muscular dystrophy. *Clinical Nursing Research*, *15*(2), 86–104. doi:10.1177/1054773805285706
- Bradbury-Jones, C., Irvine, F., & Sambrook, S. (2010). Phenomenology and participant feedback: Convention or contention? *Nurse Researcher*, *17*(2), 25–33.
- Chouinard, M. C., Gagnon, C., Laberge, L., Tremblay, C., Cote, C., Leclerc, N., & Mathieu, J. (2009). The potential of disease management for neuromuscular hereditary disorders. *Rehabilitation Nursing*, *34*, 118–126.
- Cup, E. H., Kinébanian, A., Satink, T., Pieterse, A. J., Hendricks, H. T., Oostendorp, R. A., ... van Engelen, B. G. (2011). Living with myotonic dystrophy; what can be learned from couples? A qualitative study. *BMC Neurology*, *11*, 86.
- de Die-Smulders, C. E., Höweler, C. J., Thijs, C., Mirandolle, J. F., Anten, H. B., Smeets, H. J., ... Geraedts, J. P. (1998). Age and causes of death in adult-onset myotonic dystrophy. *Brain*, *121*, 1557–1563.
- Delaporte, C. (1998). Personality patterns in patients with myotonic dystrophy. *Archives of Neurology*, *55*(5), 635–640.
- Faulkner, C. L., & Kingston, H. M. (1998). Knowledge, views, and experience of 25 women with myotonic dystrophy. *Journal of Medical Genetics*, *35*(12), 1020–1025.
- Gagnon, C., Mathieu, J., & Noreau, L. (2007). Life habits in myotonic dystrophy type 1. *Journal of Rehabilitation Medicine*, *39*(7), 560–566.
- Garrett, J. M., DuBose, T. D. Jr., Jackson, J. E., & Norman, J. R. (1969). Esophageal and pulmonary disturbances in myotonia dystrophica. *Archives of Internal Medicine*, *123*(1), 26–32.
- Genoe, M. R., & Dupuis, S. L. (2013). Picturing leisure: Using photovoice to understand the experience of leisure and dementia. *Qualitative Report*, *18*, 1–21.
- Goodyear-Smith, F., & Buetow, S. (2001). Power issues in the doctor-patient relationship. *Health Care Analysis*, *9*, 449–462.
- Harper, P. S. (2001). *Myotonic dystrophy* (3rd ed.). London, England: Harcourt.
- Heatwole, C., Bode, R., Johnson, N., Quinn, C., Martens, W., McDermott, M. P., ... Moxley, R. III. (2012). Patient-reported

- impact of symptoms in myotonic dystrophy type 1 (PRISM-1). *Neurology*, 79(4), 348–357.
- Heesen, C., Köpke, S., Richter, T., & Kasper, J. (2007). Shared decision making and self-management in multiple sclerosis—A consequence of evidence. *Journal of Neurology*, 254(Suppl. 2), 116–121.
- Hill, J. (1997). Patient satisfaction in a nurse-led rheumatology clinic. *Journal of Advanced Nursing*, 25(2), 347–354.
- Jurkowski, J. M., & Paul-Ward, A. (2007). Photovoice with vulnerable populations: Addressing disparities in health promotion among people with intellectual disabilities. *Health Promotion Practice*, 8(4), 358–365.
- Laberge, L., Mathieu, J., Auclair, J., Gagnon, É., Noreau, L., & Gagnon, C. (2013). Clinical, psychosocial, and central correlates of quality of life in myotonic dystrophy type 1 patients. *European Neurology*, 70(5–6), 308–315.
- LaDonna, K. A., Koopman, W. J., Ray, S. L., & Venance, S. L. (in press). Hard to swallow: A phenomenological exploration of caregivers for individuals with myotonic dystrophy (myotonic dystrophy) and dysphagia. *Journal of Neuroscience Nursing*.
- Levin, T., Scott, B. M., Borders, B., Hart, K., Lee, J., & Decanini, A. (2007). Aphasia Talks: Photography as a means of communication, self-expression, and empowerment in persons with aphasia. *Topics in Stroke Rehabilitation*, 14(1), 72–84.
- Lorenz, L. S., & Kolb, B. (2009). Involving the public through participatory visual research methods. *Health Expectations*, 12(3), 262–274.
- Martinez-Martin, P., Rodriguez-Blazquez, C., Kurtis, M. M., Chaudhuri, K. R., & NMSS Validation Group. (2011). The impact of non-motor symptoms on health-related quality of life of patients with Parkinson's disease. *Movement Disorders*, 26(3), 399–406.
- Meola, G., & Sansone, V. (2007). Cerebral involvement in myotonic dystrophies. *Muscle and Nerve*, 36(3), 294–306.
- Meola, G., Sansone, V., Perani, D., Scarone, S., Cappa, S., Dragoni, C., ... Moxley, R. T. (2003). Executive dysfunction and avoidant personality trait in myotonic dystrophy type 1 (DM-1) and in proximal myotonic myopathy (PROMM/DM-2). *Neuromuscular Disorders*, 13(10), 813–821.
- Miles, M. B., Huberman, A. M., & Saldana, J. (2014). *Qualitative data analysis: A methods source book* (3rd ed.). Thousand Oaks, CA: Sage.
- Nätterlund, B., Sjöden, P. O., & Ahlström, G. (2001). The illness experience of adult persons with muscular dystrophy. *Disability and Rehabilitation*, 23(17), 788–798.
- Nierse, C. J., Abma, T. A., Horemans, A. M., & van Engelen, B. G. (2013). Research priorities of patients with neuromuscular disease. *Disability and Rehabilitation*, 35(5), 405–412.
- Schipper, K., Dauwese, L., Hendriks, A., Leedekerken, J. W., & Abma, T. A. (2014). Living with Parkinson's disease: Priorities for research suggested by patients. *Parkinsonism and Related Disorders*, 20(8), 862–866.
- Sistiaga, A., Urretta, I., Jodar, M., Cobo, A. M., Emparanza, J., Otaegui, D., ... López de Munain, A. (2010). Cognitive/personality pattern and triplet expansion size in adult myotonic dystrophy type 1 (DM1): CTG repeats, cognition and personality in DM1. *Psychological Medicine*, 40(3), 487–495.
- Stromberg, A., Martensson, J., Fridlund, B., Levin, L. A., Karlsson, J. E., & Dahlstrom, U. (2003). Nurse-led heart failure clinics improve survival and self-care behaviour in patients with heart failure—results from a prospective, randomised trial. *European Heart Journal*, 24(11), 1014–1023.
- Teunissen, T., Visse, M., de Boer, P., & Abma, T. A. (2013). Patient issues in health research and quality of care: An inventory and data synthesis. *Health Expectations*, 16(4), 308–322.
- Wang, C., & Burris, M. A. (1994). Empowerment through photo novella: Portraits of participation. *Health Education Quarterly*, 21(2), 171–186.
- Wang, C., & Burris, M. A. (1997). Photovoice: Concept, methodology, and use for participatory needs assessment. *Health Education and Behavior*, 24(3), 369–387.
- Wiersma, E. C. (2011). Using Photovoice with people with early-stage Alzheimer's disease: A discussion of methodology. *Dementia*, 10(2), 203–216.